



4191-02U

SOCIAL SECURITY ADMINISTRATION

20 CFR Part 404

[Docket No. SSA-2010-0055]

RIN 0960-AF88

Revised Medical Criteria for Evaluating Hematological Disorders

AGENCY: Social Security Administration.

ACTION: Final rules.

SUMMARY: We are revising the criteria in the Listing of Impairments (listings) that we use to evaluate cases involving hematological disorders in adults and children under titles II and XVI of the Social Security Act (Act). These revisions reflect our adjudicative experience, advances in medical knowledge, diagnosis, and treatment, and public comments we received in response to a Notice of Proposed Rulemaking (NPRM).

DATES: These rules are effective [Insert date 30 days after date of publication in the FEDERAL REGISTER].

FOR FURTHER INFORMATION CONTACT: Cheryl Williams, Office of Medical Policy, Social Security Administration, 6401 Security Boulevard, Baltimore, Maryland 21235-6401, (410) 965-1020. For information on eligibility or filing for benefits, call our

national toll-free number, 1-800-772-1213, or TTY 1-800-325-0778, or visit our Internet Website, Social Security Online, at <http://www.socialsecurity.gov>.

SUPPLEMENTARY INFORMATION:

Background

We are revising and making final the rules for evaluating hematological disorders that we proposed in an NPRM published in the Federal Register on November 19, 2013 at 78 FR 69324. Even though these rules will not go into effect until 30 days after publication of this document, for clarity, we refer to them in this preamble as the “final” rules. We refer to the rules in effect prior to that time as the “prior” rules.

In the preamble to the NPRM, we discussed the revisions we proposed for the hematological disorders body system. Since we are mostly adopting those revisions as we proposed them, we are not repeating that information here. Interested readers may refer to the preamble to the NPRM for this information, available at <http://www.regulations.gov>.

We are making several changes in these final rules from the NPRM based upon some of the public comments we received. We explain these changes below in the “Summary of Public Comments on the NPRM” section of this preamble.

Why are we revising the listings for hematological disorders?

We developed these final rules as part of our ongoing review of the listings. When

we last comprehensively revised the listings for the hematological disorders body system in final rules published on December 6, 1985, we indicated in the preamble to those rules that we would carefully monitor these listings to ensure that they continue to meet program purposes, and that we would update them if warranted.¹

Summary of Public Comments on the NPRM

In the NPRM, we provided the public with a 60-day comment period that ended on January 21, 2014. We received 32 comments. The commenters included advocacy groups, a national group representing disability examiners in the State agencies that make disability determinations for us, State agencies, groups representing medical practitioners, and individual members of the public. A number of the letters provided identical comments and recommendations.

We carefully considered all of the significant comments relevant to this rulemaking. We condensed and summarized the comments below. We presented the commenters' concerns and suggestions and responded to all significant issues that were within the scope of these rules. We provide our reasons for adopting or not adopting the recommendations in our responses below.

General Comments

¹ See 50 FR 50068. We published some revisions to the hematological body system on April 24, 2002, and November 15, 2004. See 67 FR 20018 and 69 FR 67017 (corrected at 70 FR 15227). These revisions were not comprehensive; they addressed only specific listings.

Comment: One commenter recommended that we review the medical criteria in the listings for evaluating hematological disorders every five years to ensure they reflect the latest advances in treatment and clinical practice. The commenter thought it especially important that we review ongoing clinical trials and published reports regarding advances in genetic testing and the clinical use of new blood derivatives and biologics.

Response: While we agree with the commenter that it is important to keep abreast of advances in treatment and clinical practice for hematological disorders, we have not made any changes to our proposed listings as a result of this comment. As mentioned above, we will monitor the final rules to ensure they still meet our program purposes. While doing this, we will consider whether we need to revise the rules to reflect advances in medical knowledge and clinical practice.²

Comment: Several commenters expressed concern that people with hematological disorders may be disabled but their impairments do not satisfy the specific medical criteria in the listings. The commenters said these people may have periods of relative functional ability punctuated by unpredictable and episodic complications that result in an inability to work. They believed such complications do not necessarily have to be prolonged or frequent to be disabling and to result in loss of employment, failure in

² We have made it a priority to ensure that we keep the listings up to date and to report our progress. For example, see SSA's Annual Performance Plan for Fiscal Year 2015, Revised Performance Plan for Fiscal Year 2014, and Annual Performance Report for Fiscal Year 2013 available at <http://www.ssa.gov/agency/performance/2015/FY2015-APP-APR.pdf>.

school, or other major disruptions in the person's life.

Response: We agree that many of these final listings have specific medical criteria. Some people with hematological disorders may have complications that do not occur with the severity or frequency that these listings require. We believe the functional criteria in our final rules address commenters' concerns by providing criteria that may permit a finding of disability at the listing step of the sequential evaluation process in people who suffer repeated complications of their impairments, but who may not be continually restricted in their functioning between complications. For example, our intent in new functional listing 7.18 for adults, and in our functional equivalence rules for children, is to evaluate impairments that are difficult to assess in strict medical terms. We can use the functional criteria in listing 7.18, as well as our functional equivalence rules in claims for childhood disability under the Supplemental Security Income (SSI) program, to evaluate claims filed by people who become ill and improve, but become ill again, either with the same complications of their hematological disorders or with different ones.

Comment: One commenter recommended we add a criterion in these final rules requiring compliance with prescribed therapy.

Response: We did not adopt the commenter's recommendation because we believe our adjudicators can establish the relevance of a person's noncompliance under our current rules and current operating instructions regarding failure to follow prescribed

treatment.³ Under our policy, we must assess a person's noncompliance on an individual basis because the person may have good cause for not following prescribed treatment. Good cause may include concern about the cost or adverse effects of treatment, lack of access to treatment, religious beliefs, or other situations. We also provide information to our adjudicators in final sections 7.00H and 107.00G on how to consider whether a person is receiving or following treatment.

Comment: Another commenter recommended that the final listings consider the cost of medication for treating hematological disorders before denying children's disability claims.

Response: We did not adopt the comment because, as just indicated, we will consider on an individual case basis whether a person, including a child, can afford, or has access to, medically necessary treatments.

Comment: Some commenters objected to our use of hospitalization as a criterion in several final listings for determining listing-level severity of a person's hematological disorder. These listings require hospitalization at least three times within a 12-month period, with each hospitalization occurring at least 30 days apart. The commenters believed health insurers and hospitals are actively trying to reduce hospital admissions, which may prevent some disabled people from receiving benefits. One commenter

³ See 20 CFR 404.1530 and 416.930; also see Social Security Ruling 82-59: Titles II and XVI: Failure to Follow Prescribed Treatment available at: http://www.socialsecurity.gov/OP_Home/rulings/di/02/SSR82-59-di-02.html); and also see DI 23010 Failure to Follow Prescribed Treatment—Procedures, Program Operations Manual System (POMS), available at: <https://secure.ssa.gov/apps10/poms.nsf/lnx/0423010000>.

thought that discrimination and a lack of uniformity of treatment protocols among communities and hospitals could also affect decisions regarding hospitalization. The commenters recommended we delete the hospitalization requirement or require fewer than three hospitalizations in a 12-month period. Some commenters also recommended we consider the frequency of outpatient visits as a measure of listing-level severity.

Response: We decided to retain the hospitalization criterion because our intent in these final listings is to reflect criteria that result in an inability to perform any gainful activity, which can be demonstrated by a need for a level of care beyond more conventional treatments for hematological disorders. We believe the hospitalization criterion is an advantage to people who apply for disability benefits because it provides another way for us to find them disabled at the listing step.

We want to assure the commenters that we are able to evaluate hematological disorders resulting in fewer than three hospitalizations in a consecutive 12-month period under the criteria in final listing 7.18 for adults, the functional equivalence rules for children, or at other steps in our sequential evaluation process. For example, the criteria in listing 7.18 evaluate the functional impact of the person's impairment in the broad areas of activities of daily living, social functioning, and concentration, persistence, or pace, including the functional impact of treatment such as repeated outpatient visits for complications. We are also able to evaluate hematological disorders that are "severe" but do not meet or equal any listing under the final steps of the sequential evaluation process.

Comment: One commenter expressed concern that people with hematological disorders may have complications and co-occurring conditions for years, but their impairments never result in hospitalization. This commenter was also concerned that our adjudicators may not know about many of the hematological disorders, their effects, and how to recognize them.

Response: As previously discussed, we believe the functional criteria in listing 7.18 and our childhood functional equivalence criteria under the SSI program will help us determine disability appropriately for people whose hematological disorders result in fewer than three hospitalizations in a 12-month period. These criteria also cover people who have never been hospitalized.

With regard to the commenter's concerns about adjudicators' knowledge of hematological disorders, the introductory text and listings provide common examples of hematological disorders and describe their complications. However, we do not think it is practical or necessary to list all hematological disorders and their complications. Instead, as we do with respect to other changes in our listings, we plan to provide instructions and training to our adjudicators. These instructions and the training will help our adjudicators recognize less common examples of hematological disorders and their associated complications and functional limitations.

Comment: One commenter believed the requirement that the hospitalization must last at least 48 hours seems to be "arbitrary" and not based on scientific or medical

standards. The commenter thought it would be just as appropriate for us to require the hospitalization to last at least 24 hours, as listings in some other body systems require.

Response: We disagree with the commenter that we should require hospital stays of at least 24 hours. As we noted in the preamble of the NPRM, the 48-hour criterion more clearly defines our intent in prior listing 7.05B for an “extended hospitalization.”⁴ This criterion is more detailed than in the prior listing, but it is not stricter. We believe the scientific and medical literature shows that many people hospitalized for serious complications of hematological disorders are included in the 48-hour criterion, and that this criterion can help identify an impairment of listing-level severity.

In sickle cell disease, for instance, a 2008 study found 63 percent of children hospitalized for pain crises had hospital stays of at least 4 days, not counting time in the emergency department.⁵ Similarly, a 2004 study of children hospitalized for sickle cell disease complications other than strokes reported a median hospital stay of 3 days⁶; children with strokes had a median hospital stay of 6 days. A 2010 study of adults and children with sickle cell complications reported an average initial hospital stay of 5.6 days.⁷ Children in the 2008 study with long hospital stays tended to have high pain scores, pain in multiple body sites, co-occurring complications, and a need for extensive treatment.

⁴ 78 FR at 69328.

⁵ Rogovik, A., *et al.*, Admission and length of stay due to painful vasoocclusive crisis in children, The American Journal of Emergency Medicine, Sep;27(7), 797-801 (2008).

⁶ Fullerton, H., *et al.*, Declining stroke rates in California children with sickle cell disease, Blood, Jul;104(2), 336-339 (2004).

⁷ Brousseau, D., *et al.*, Acute care utilization and rehospitalizations for sickle cell disease, Journal of the American Medical Association, Apr;303(13), 1288-1294 (2010).

In hemophilia, a study published in 2011 of Texas patients with bleeding episodes reported a median hospital stay of 4 days.⁸ A 2005 study of patients with potentially life-threatening bleeds in the iliopsoas muscle reported a median hospital stay of 4.8 days.⁹ Hospital stays may be longer for iliopsoas bleeds in hemophiliacs with “inhibitors” (replacement factor alloantibodies).¹⁰ Generally, hemophiliacs with inhibitors may require more extensive treatment than those without inhibitors because their bleeding episodes often are resistant to standard treatments.¹¹

The study findings described above are consistent with our adjudicative experience that many claimants with listing-level hematological disorders satisfy the 48-hour criterion because their complications are difficult to treat and recoveries are prolonged.¹² On the other hand, we believe requiring the hospitalization to last at least 24 hours would not be an accurate predictor of impairment severity because this criterion would include people who recover relatively quickly and satisfactorily with standard treatments. These hospitalizations include people hospitalized only overnight, for example, to receive extra fluids after treatment in the emergency department, and those kept for observation after surgery. In this regard, a 24-hour criterion would not reflect our

⁸ Mirchandani, G.G., et al., Surveillance of bleeding disorders, Texas, 2007, American Journal of Preventive Medicine, Dec;41(6 Suppl 4), S354-359 (2011).

⁹ Balkan, C., et al., Iliopsoas haemorrhage in patients with haemophilia: Results of one centre, *Haemophilia*, Sep;11(5), 463-467 (2005).

¹⁰ Ashrani, A.A., et al., Iliopsoas haemorrhage in patients with bleeding disorders—experience from one centre, *Haemophilia*, Nov;9(6), 721-726 (2003).

¹¹ Soucie, J.M., et al., Home-based factor infusion therapy and hospitalization for bleeding complications among males with haemophilia, *Mar;7(2)*, 198-206 (2001).

¹² The study findings only expand on, and confirm, the data in the studies we cited in the NPRM. They do not change either the methodology in the listing or any substantive criteria in it.

intent that the listing be used to evaluate impairments at the listing level, which require treatment beyond the usual course of treatment for the hematological disorder.

Comment: One commenter questioned our use of the term “disorders of hemostasis” in the introductory text and the listings. The commenter noted that the medical community usually refers to the grouping of clotting and bleeding disorders as “disorders of thrombosis and hemostasis.”

Response: We adopted the comment and modified the listings accordingly.

Comment: Some commenters suggested minor editorial changes in the introductory text, such as a comment asking us to indicate that the examples of complications of hematological disorders in section 7.00C, section 7.00D, and other final sections are not all-inclusive.

Response: We made these minor editorial changes for clarity and consistency; none were substantive.

Sections 7.00B and 107.00B--What evidence do we need to document that you have a hematological disorder?

Comment: Some commenters expressed concern over the requirement in proposed sections 7.00B and 107.00B that laboratory reports of definitive tests

establishing hematological disorders have a physician's signature. These commenters thought this requirement too difficult or burdensome for some claimants because it may require them to obtain additional medical evidence. These commenters said it is not the usual practice for the overseeing physician in a laboratory to sign laboratory reports of definitive tests. They recommended we accept reports signed by treating physicians or other physicians if these reports state that the definitive hematological evidence is present in the medical records. They also believed we should accept a physician's statement that a person has a hematological disorder, even if the definitive hematological evidence is not present in the medical records.

Response: We did not adopt the comments. Under our policy, evidence establishing a medically determinable impairment (MDI) must be appropriately developed. To develop this evidence appropriately, it must come from acceptable medical sources, that is, medical or osteopathic doctors.¹³ A doctor's signature on a definitive laboratory test establishing that the person has a hematological disorder confirms the evidence came from an acceptable medical source, and we do not need to develop the evidence further to establish an MDI. In situations in which a doctor did not sign the definitive laboratory test, we will continue to develop the evidence. Final sections 7.00B and 107.00B provide examples of additional evidence we may obtain from doctors to establish the MDI, and we believe these examples are comparable to what the commenters recommended. Consequently, final sections 7.00B and 107.00B clarify how we develop evidence establishing the MDI; they do not add new requirements.

¹³ See 20 CFR 404.1513(a) and 416.913(a).

Sections 7.00C and 107.00C--What are hemolytic anemias, and how do we evaluate them under 7.05 and 107.05?

Comment: One commenter pointed out that hemolytic anemias are sometimes acquired conditions.

Response: We adopted this comment and revised final sections 7.00C1 and 107.00C1 to provide examples of acquired hemolytic anemias. We made similar changes in final sections 7.00D1, 107.00D1, 7.00E1, and 107.00E1. We also provided examples of acquired disorders of thrombosis and hemostasis, as well as disorders of bone marrow failure.

Comment: A commenter recommended that we add hereditary spherocytosis to the list of common examples of hemolytic anemias in adults. The commenter also suggested that we add paroxysmal nocturnal hemoglobinuria to the list of examples.

Response: We adopted this recommendation and added hereditary spherocytosis to the list in 7.00C1. We also added hereditary spherocytosis to the list of common examples of hemolytic anemias in children in 107.00C1 to make the child listings consistent with the adult listings.

We did not adopt the commenter's recommendation that we add paroxysmal

nocturnal hemoglobinuria to the list of examples. Although we evaluate paroxysmal nocturnal hemoglobinuria under the hematological disorders listings, it is a very rare disorder. We provide only examples of common hemolytic anemias in the listings because we do not believe it is practical or necessary to name all of the hematological disorders we evaluate under this body system. We plan to provide information to our adjudicators about less common examples of hematological disorders, such as paroxysmal nocturnal hemoglobinuria, through training and operating instructions.

Comment: We received many comments expressing concern over our exclusion in proposed sections 7.00C4 and 107.00C4 of prophylactic red blood cell (RBC) transfusions to prevent stroke in people with sickle cell disease. Some of these commenters recommended that we delete the statement in proposed section 7.00C4 that we do not consider prophylactic RBC transfusions for sickle cell disease to be of equal medical significance to transfusion-dependent thalassemia.¹⁴ They said people with sickle cell disease who require prophylactic RBC transfusions are usually chronically ill, and they cited articles in the current medical literature to support their views. Another commenter believed final sections 7.00C4 and 107.00C4 needed more information to help adjudicators determine whether the need for RBC transfusions will be life-long.

The commenters also believe people with sickle cell disease who receive prophylactic RBC transfusion to prevent stroke may be more severely impaired than people with transfusion-dependent beta thalassemia major because they have a far greater

¹⁴ 78 FR at 69333.

burden of cerebrovascular disease and intellectual and physical impairment. Additionally, a comment from a national advocacy group for physicians in pediatric hematology and oncology said its membership now considers sickle cell disease with stroke to be a transfusion-dependent disorder like thalassemia because of the risk of recurrent strokes if prophylactic RBC transfusion stops.

Response: We do not agree that treatment with prophylactic RBC transfusions alone should reflect a listing-level impairment in sickle cell disease and have not adopted the commenters' recommendations. Under the Act, we cannot find that a person is disabled based on the risk of a complication occurring in the future, as, for example, when transfusion therapy is effective and the person has not experienced a stroke.

However, we agree that people with sickle cell disease are chronically sick. We added language to final sections 7.00C4 and 107.00C4 that directs evaluation under listings 11.00, 111.00, 12.00, and 112.00 if a claimant has had a stroke. We also added language in final sections 7.00C4 and 107.00C4 explaining that we will consider functional limitations associated with chronic RBC transfusions under final listing 7.18 for adults, the functional equivalence rules for children, as well as the listings for any affected body systems. The additional language also addresses complications resulting from chronic RBC transfusion, such as iron overload.

We also deleted the term "transfusion-dependent" in the final sections 7.00C4, 107.00C4, 7.00E3, and 107.00E3 because comments demonstrated to us that this term

may confuse adjudicators. We made a corresponding change in final listings 7.05D, 107.05D, 7.10B, and 107.10B. Instead, we use the phrase, “requiring RBC transfusions at least once every 6 weeks to maintain life.” We believe this phrase is more descriptive of our intent in these final rules, which is that listing-level severity for hematological disorders requires treatment with RBC transfusions that are life-saving in nature and life-long in need. Moreover, we are confident our adjudicators will understand the requirement that the RBC transfusions must be “life-long,” as reflected in the ultimately fatal nature of beta thalassemia major and myelodysplastic syndrome if this treatment is withdrawn.

Sections 7.00D and 107.00D--What are disorders of thrombosis and hemostasis, and how do we evaluate them under 7.08 and 107.08?

Comment: A commenter noted that the future development of new treatments for hemophilia may make the term “factor infusions” less relevant.

Response: We adopted the comment and use the term “clotting-factor proteins” in final sections 7.00D2 and 107.00D2, instead of the term “factor infusions.”

Comment: A commenter stated that the language in proposed sections 7.00D2 and 107.00D2 was vague and did not make it clear that these sections included any surgery.

Response: We revised final sections 7.00D2 and 107.00D2 to state explicitly that

we consider all surgeries in people with disorders of thrombosis or hemostasis to be complications of their disorders if they needed treatment with clotting-factor proteins or anticoagulant medications to control bleeding or coagulation in connection with the surgery.

Sections 7.00I and 107.00H--How do we evaluate episodic events in hematological disorders?

Comment: Some commenters thought proposed sections 7.00I and 107.00I could imply that the consecutive 12-month period required for episodic events could not include the months before a person files a disability claim, or the months before the person's alleged onset date of disability.

Response: In response to these comments, we added language to clarify the guidance in final sections 7.00I and 107.00I.

Listings 7.05 and 107.05--Hemolytic anemias, including sickle cell disease, thalassemia, and their variants

Comment: Several commenters expressed concern about the criterion in proposed listings 7.05A and 107.05A requiring at least six pain crises treated with parenteral narcotic medications within a 12-month period and occurring at least 30 days apart. These commenters believed this criterion is too restrictive, particularly for evaluating

sickle cell disease. They believed that recent scientific and medical literature points to three pain crises requiring parenteral narcotic medication within a 12-month period as a more appropriate standard.

Some commenters also noted that pain crises treated with only oral narcotic medications may be severe enough to disrupt a person's life for days or weeks. These commenters believed such pain crises greatly impair a person's mobility, self-care, and mental capacity, and they noted that there can be long-term, cumulative tissue and organ damage associated with the crises. A national advocacy group for persons with hematological disorders recommended we consider the daily use of oral opioids as a criterion for listing-level severity. The group provided a suggested revision to final listing 7.05A that considered a person disabled if he or she required daily oral opioids for chronic pain for a period of at least 30 consecutive days, at least three times within a 12-month period.

Response: We did not adopt these comments because we believe final listings 7.05A and 107.05A provide objective criteria that are more descriptive of our intent and more specific to listing-level determinations than the prior listings. In addition, as we noted previously, final listing 7.18 provides criteria to evaluate claims from individuals whose impairments do not satisfy the medical criteria in final listing 7.05A, but whose impairments result in functional limitations that meet the criteria of listing 7.18. These effects may include chronic pain and other complications, as well as a frequent need for oral narcotic medication or other treatments that may cause negative side effects. Some

people with sickle cell disease or other hemolytic anemia may have impairments that are less than listing-level severity, but may still be disabling. We can evaluate these impairments through the steps of our sequential evaluation process after the listing step.

Comment: One commenter noted that a person hospitalized for pain crises may receive treatments other than parenteral narcotic medication, such as local or regional anesthetic blocks. The commenter believed pain crises requiring such treatments also result in functional impairments and are indicative of pain severity, but were not reflected in proposed listings 7.05A and 107.05A.

Response: While it is true final listings 7.05A and 107.05A do not specify these other treatments, we did not adopt this comment because we are able to evaluate hospitalizations for pain crises treated with other treatments under final listings 7.05B and 107.05B, or we can evaluate the functional impairments described by the commenter under final listing 7.18, or the functional equivalence rules for childhood disability claims under the SSI program.

Comment: One commenter agreed with the requirement in listings 7.05B and 107.05B that a hospitalization should last at least 48 hours, but recommended that this criterion not include hours spent in the hospital emergency department immediately before the hospitalization. The commenter said hospitals may not always document patients' arrival times in their emergency departments and times of discharge to inpatient units.

Response: We did not adopt the commenter's recommendation because our adjudicative experience shows that hospitals document these times in the great majority of cases.

Comment: A commenter suggested we count the hours a person receives treatment in a comprehensive sickle cell disease center under our requirement in final listings 7.05B and 107.05B that hospitalizations for complications of hemolytic anemias last at least 48 hours. We received a similar comment regarding comprehensive hemophilia treatment centers.

Response: We adopted these comments. We explain in final sections 7.00C2 and 107.00C2 that we will count the hours the person receives treatment in a comprehensive sickle cell disease center if the treatment is comparable to the treatment provided in a hospital emergency department. We also revised final listings 7.08 and 107.08 and final sections 7.00D2 and 107.00D2 in response to the comment regarding comprehensive hemophilia treatment centers.

Comment: One commenter believed the requirement in proposed listings 7.05B and 107.05B for three hospitalizations within a 12-month period is too restrictive because it applies only to a subset of people with sickle cell disease who the commenter described as "high-risk" patients. The commenter believed we should consider a person with sickle cell disease to be disabled if he or she has any of the complications described in final

sections 7.00C2 and 107.00C2 because this person needs continual follow-up and monitoring regardless of hospitalization.

Response: While we appreciate the commenter's concerns—and we agree that people with sickle cell disease have serious impairments if they have any of the complications described in final sections 7.00C2 and 107.00C2—we did not adopt the comment. We can evaluate these claimants' impairments under any appropriate listing in the affected body system, or at the steps of our sequential evaluation process after the listing step, if they do not meet or medically equal the criteria in listings 7.05B and 107.05B.

Comment: We received a comment recommending we add guidance to the listings that explains to adjudicators they can use hematocrit readings under final listings 7.05C and 107.05C if a person's case record does not include hemoglobin measurements. The commenter was concerned adjudicators might misinterpret the listings to mean they cannot use hematocrit readings under any circumstances.

Response: We did not adopt this recommendation. These final listings require hemoglobin measurements at 7.0 grams per deciliter (g/dL) or less, occurring at least three times within a 12-month period with at least 30 days between measurements. In the great majority of cases, our adjudicative experience shows a person's case record provides both hemoglobin measurements and hematocrit readings. Moreover, we are confident that our adjudicators understand they can use comparable hematocrit levels to

medically equal the listings if hemoglobin measurements are not available. The final listings do not provide substantive instructions to our adjudicators for determining such equivalence because we can better provide this information through operating instructions and training.

Comment: Two commenters questioned whether we should use hemoglobin measurements at all. One commenter said the science and the medical communities have not established a critical threshold for hemoglobin for determining disability. The other commenter said disability depends on factors besides hemoglobin level, such as the duration of anemia, the bone marrow's response, and associated cardiovascular or other organ dysfunction. For children, this commenter said we should also consider amount of fatigue, inability to concentrate, problems with executive function, and memory deficiencies.

Response: We did not adopt these comments because we believe this criterion is reasonable for quickly identifying people whose hemolytic anemias are clearly disabling, and whose claims should be allowed at the listing step. Hemoglobin at 7.0 g/dL or less can result in an abnormal heartbeat, shortness of breath with mild exertion, significant fatigue, and other very serious complications. Given these complications, we believe the criteria in the final listings reflect a persistence of very low hemoglobin that can prevent an adult from working, or prevent a child from functioning independently, appropriately, and effectively in an age-appropriate manner.

Comment: A commenter noted that people with sickle cell disease and a history of frequent pain crises or acute chest syndrome may be receiving prophylactic RBC transfusions to alleviate these complications and are not likely to have hemoglobin measurements of 7.0 g/dL. The commenter recommended that listings 7.05C and 107.05C allow for a finding of disability for people who receive prophylactic RBC transfusions for these complications.

Response: We did not adopt the comment because the intent of the hemoglobin finding in final listings 7.05C and 107.05C is to provide a faster way for us to determine listing-level disability without needing to consider a person's specific complications.

Comment: The same commenter also thought that adjudicators will have difficulty identifying hemoglobin measurements of 7.0 g/dL among potentially hundreds of measurements in a person's case record.

Response: We did not adopt this comment. We agree that a person's case record may provide many hemoglobin measurements; however, our adjudicators are accustomed to evaluating such evidence.

Listing 7.18--Repeated complications of hematological disorders

Comment: One commenter suggested that we add "chronic skin ulcers" to the examples of complications in final listing 7.18.

Response: We did not adopt this comment. Both the proposed rules and these final rules include skin ulcers as a possible complication that we will evaluate under listing 7.18. However, skin ulcers and other complications we evaluate under the listing do not have to be chronic. We explain in final section 7.00G2 that a person's complications do not have to be the same each time, but can vary. A person could have skin ulcers once and may satisfy this criterion in the listing if he or she also has other complications during the period we are considering in connection with the application.

Comment: A commenter suggested we include chronic, non-vascular necrosis-related low back pain in final listing 7.18 as a complication of a hematological disorder. The commenter also suggested that listing 7.18 take into consideration pain resulting from prolonged periods of standing or physical activity in people who have chronic pain from a hematological disorder such as sickle cell disease.

Response: We did not believe it was necessary to adopt the commenter's suggestions. The pain resulting from repeated complications of hematological disorders that listing 7.18 requires can include the chronic pain the commenter describes.

Comment: One commenter believed that it is important for adjudicators to give appropriate weight to evaluations by nurses, social workers, and physical therapists when determining a person's functional limitations under final listing 7.18.

Response: We agree that such sources can provide important information to show the severity of a person's impairment and how it affects his or her ability to work, and we currently provide guidance to our adjudicators in our regulations for considering this evidence and who may provide it.¹⁵

Listing 107.08--Disorders of hemostasis, including hemophilia and thrombocytopenia

Comment: A commenter believed proposed listing 107.08 did not recognize the developmental and functional impact that disability has on children and should reflect a need for frequent medical intervention, not only hospitalizations. The commenter stated that repeated hospitalizations and frequent outpatient medical treatment affect children much more profoundly than adults.

Response: We did not adopt the commenter's recommendation because we can evaluate the functional and developmental impact of a child's frequent medical treatment under our functional equivalence rules. Under these rules, we evaluate how independently, appropriately, and effectively the child functions compared to children of the same age who do not have a hematological disorder. This evaluation includes assessing what activities the child cannot do, has difficulty doing, or is restricted from doing because of the interactive and cumulative effects of his or her disorder and medical care.

¹⁵ See 20 CFR 404.1513(d), 20 CFR 416.913(d), and Social Security Ruling 06-03p: Titles II and XVI: Considering Opinions and Other Evidence from Sources Who Are Not "Acceptable Medical Sources" in Disability Claims, 71 FR 45593 (2006) (also available at: http://www.ssa.gov/OP_Home/rulings/di/01/SSR2006-03-di-01.html).

Listing 107.10--Disorders of bone marrow failure, including myelodysplastic syndromes, aplastic anemia, granulocytopenia, and myelofibrosis

Comment: A commenter stated that the requirement in 107.10A for three hospitalizations within a 12-month period may be too restrictive for children because “impairment can be severe in a child” following a single hospitalization.

Response: We did not modify the proposed listing as a result of this comment. We believe the hospitalization criterion for disorders of bone marrow failure is an advantage to children and adults who apply for disability benefits because it provides another way we may find them disabled at the listing step. Additionally, the child functional equivalence rules help us evaluate SSI claims filed by children whose hematological disorders result in fewer than three hospitalizations in a 12-month period.

What is our authority to make rules and set procedures for determining whether a person is disabled under the statutory definition?

Under the Act, we have authority to make rules and regulations and to establish necessary and appropriate procedures to carry out such provisions.¹⁶

How long will these final rules be in effect?

¹⁶ See sections 205(a), 702(a)(5), and 1631(d)(1).

These final rules will be in effect for 5 years after their effective date, unless we extend them. We will continue to monitor these rules to ensure that they continue to meet program purposes, and may revise them before the end of the 5-year period if warranted.

REGULATORY PROCEDURES

Executive Order 12866, as Supplemented by Executive Order 13563

We consulted with the Office of Management and Budget (OMB) and determined that these final rules meet the requirements for a significant regulatory action under Executive Order 12866, as supplemented by Executive Order 13563 and was reviewed by OMB.

Regulatory Flexibility Act

We certify that these final rules will not have a significant economic impact on a substantial number of small entities because they affect only individuals. Therefore, the Regulatory Flexibility Act, as amended, does not require us to prepare a regulatory flexibility analysis.

Paperwork Reduction Act

These final rules do not impose new or affect any existing reporting or recordkeeping requirements and are not subject to OMB clearance.

(Catalog of Federal Domestic Assistance Program Nos. 96.001, Social Security—Disability Insurance; 96.002, Social Security—Retirement Insurance; 96.004, Social Security—Survivors Insurance; and 96.006, Supplemental Security Income).

List of Subjects in 20 CFR Part 404

Administrative practice and procedure, Blind, Disability benefits, Old-age, Survivors and Disability Insurance, Reporting and recordkeeping requirements, Social Security.

Dated: April 10, 2015.

Carolyn W. Colvin,
Acting Commissioner of Social Security.

For the reasons set out in the preamble, we are amending 20 CFR part 404, subpart P as set forth below:

Part 404—FEDERAL OLD-AGE, SURVIVORS AND DISABILITY INSURANCE
(1950-)

Subpart P—[Amended]

1. The authority citation for subpart P of part 404 continues to read as follows:

Authority: Secs. 202, 205(a)-(b) and (d)-(h), 216(i), 221(a), (i), and (j), 222(c), 223, 225, and 702(a)(5) of the Social Security Act (42 U.S.C. 402, 405(a)-(b) and (d)-(h), 416(i), 421(a), (i), and (j), 422(c), 423, 425, and 902(a)(5)); sec. 211(b), Pub. L. 104-193, 110 Stat. 2105, 2189; sec. 202, Pub. L. 108-203, 118 Stat. 509 (42 U.S.C. 902 note).

2. Amend appendix 1 to subpart P of part 404 by revising:

- a. Item 8 of the introductory text before part A;
- b. Section 7.00 of part A;
- c. Section 13.00K2c(ii) of part A;
- d. Second sentence of section 13.00K3 of part A; and
- e. Section 107.00 of part B.

The revisions read as follows:

Appendix 1 to Subpart P of Part 404—Listing of Impairments

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8. Hematological Disorders (7.00 and 107.00): May 18, 2020.

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Part A

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7.00 HEMATOLOGICAL DISORDERS

A. What hematological disorders do we evaluate under these listings?

1. We evaluate non-malignant (non-cancerous) hematological disorders, such as hemolytic anemias (7.05), disorders of thrombosis and hemostasis (7.08), and disorders of bone marrow failure (7.10). These disorders disrupt the normal development and function of white blood cells, red blood cells, platelets, and clotting-factor proteins (factors).

2. We evaluate malignant (cancerous) hematological disorders, such as lymphoma, leukemia, and multiple myeloma, under the appropriate listings in 13.00, except for lymphoma associated with human immunodeficiency virus (HIV) infection, which we

evaluate under 14.08E.

B. What evidence do we need to document that you have a hematological disorder? We need the following evidence to document that you have a hematological disorder:

1. A laboratory report of a definitive test that establishes a hematological disorder, signed by a physician; or

2. A laboratory report of a definitive test that establishes a hematological disorder that is not signed by a physician and a report from a physician that states you have the disorder; or

3. When we do not have a laboratory report of a definitive test, a persuasive report from a physician that a diagnosis of your hematological disorder was confirmed by appropriate laboratory analysis or other diagnostic method(s). To be persuasive, this report must state that you had the appropriate definitive laboratory test or tests for diagnosing your disorder and provide the results, or explain how your diagnosis was established by other diagnostic method(s) consistent with the prevailing state of medical knowledge and clinical practice.

4. We will make every reasonable effort to obtain the results of appropriate laboratory testing you have had. We will not purchase complex, costly, or invasive tests,

such as tests of clotting-factor proteins, and bone marrow aspirations.

C. What are hemolytic anemias, and how do we evaluate them under 7.05?

1. Hemolytic anemias, both congenital and acquired, are disorders that result in premature destruction of red blood cells (RBCs). Hemolytic disorders include abnormalities of hemoglobin structure (hemoglobinopathies), abnormal RBC enzyme content and function, and RBC membrane (envelope) defects that are congenital or acquired. The diagnosis of hemolytic anemia is based on hemoglobin electrophoresis or analysis of the contents of the RBC (enzymes) and membrane. Examples of congenital hemolytic anemias include sickle cell disease, thalassemia and their variants, and hereditary spherocytosis. Acquired hemolytic anemias may result from autoimmune disease (for example, systemic lupus erythematosus) or mechanical devices (for example, heart valves, intravascular patches).

2. The hospitalizations in 7.05B do not all have to be for the same complication of the hemolytic anemia. They may be for three different complications of the disorder. Examples of complications of hemolytic anemia that may result in hospitalization include osteomyelitis, painful (vaso-occlusive) crisis, pulmonary infections or infarctions, acute chest syndrome, pulmonary hypertension, chronic heart failure, gallbladder disease, hepatic (liver) failure, renal (kidney) failure, nephrotic syndrome, aplastic crisis, and stroke. We will count the hours you receive emergency treatment in a comprehensive sickle cell disease center immediately before the hospitalization if this treatment is

comparable to the treatment provided in a hospital emergency department.

3. For 7.05C, we do not require hemoglobin to be measured during a period in which you are free of pain or other symptoms of your disorder. We will accept hemoglobin measurements made while you are experiencing complications of your hemolytic anemia.

4. 7.05D refers to the most serious type of beta thalassemia major in which the bone marrow cannot produce sufficient numbers of normal RBCs to maintain life. The only available treatments for beta thalassemia major are life-long RBC transfusions (sometimes called hypertransfusion) or bone marrow transplantation. For purposes of 7.05D, we do not consider prophylactic RBC transfusions to prevent strokes or other complications in sickle cell disease and its variants to be of equal significance to life-saving RBC transfusions for beta thalassemia major. However, we will consider the functional limitations associated with prophylactic RBC transfusions and any associated side effects (for example, iron overload) under 7.18 and any affected body system(s). We will also evaluate strokes and resulting complications under 11.00 and 12.00.

D. What are disorders of thrombosis and hemostasis, and how do we evaluate them under 7.08?

1. Disorders of thrombosis and hemostasis include both clotting and bleeding disorders, and may be congenital or acquired. These disorders are characterized by

abnormalities in blood clotting that result in hypercoagulation (excessive blood clotting) or hypocoagulation (inadequate blood clotting). The diagnosis of a thrombosis or hemostasis disorder is based on evaluation of plasma clotting-factor proteins (factors) and platelets. Protein C or protein S deficiency and Factor V Leiden are examples of hypercoagulation disorders. Hemophilia, von Willebrand disease, and thrombocytopenia are examples of hypocoagulation disorders. Acquired excessive blood clotting may result from blood protein defects and acquired inadequate blood clotting (for example, acquired hemophilia A) may be associated with inhibitor autoantibodies.

2. The hospitalizations in 7.08 do not all have to be for the same complication of a disorder of thrombosis and hemostasis. They may be for three different complications of the disorder. Examples of complications that may result in hospitalization include anemias, thromboses, embolisms, and uncontrolled bleeding requiring multiple factor concentrate infusions or platelet transfusions. We will also consider any surgery that you have, even if it is not related to your hematological disorder, to be a complication of your disorder of thrombosis and hemostasis if you require treatment with clotting-factor proteins (for example, factor VIII or factor IX) or anticoagulant medication to control bleeding or coagulation in connection with your surgery. We will count the hours you receive emergency treatment in a comprehensive hemophilia treatment center immediately before the hospitalization if this treatment is comparable to the treatment provided in a hospital emergency department.

E. What are disorders of bone marrow failure, and how do we evaluate them

under 7.10?

1. Disorders of bone marrow failure may be congenital or acquired, characterized by bone marrow that does not make enough healthy RBCs, platelets, or granulocytes (specialized types of white blood cells); there may also be a combined failure of these bone marrow-produced cells. The diagnosis is based on peripheral blood smears and bone marrow aspiration or bone marrow biopsy, but not peripheral blood smears alone. Examples of these disorders are myelodysplastic syndromes, aplastic anemia, granulocytopenia, and myelofibrosis. Acquired disorders of bone marrow failure may result from viral infections, chemical exposure, or immunologic disorders.

2. The hospitalizations in 7.10A do not all have to be for the same complication of bone marrow failure. They may be for three different complications of the disorder. Examples of complications that may result in hospitalization include uncontrolled bleeding, anemia, and systemic bacterial, viral, or fungal infections.

3. For 7.10B, the requirement of life-long RBC transfusions to maintain life in myelodysplastic syndromes or aplastic anemias has the same meaning as it does for beta thalassemia major. (See 7.00C4.)

F. How do we evaluate bone marrow or stem cell transplantation under 7.17? We will consider you to be disabled for 12 months from the date of bone marrow or stem cell transplantation, or we may consider you to be disabled for a longer period if you are

experiencing any serious post-transplantation complications, such as graft-versus-host (GVH) disease, frequent infections after immunosuppressive therapy, or significant deterioration of organ systems. We do not restrict our determination of the onset of disability to the date of the transplantation in 7.17. We may establish an earlier onset date of disability due to your transplantation if evidence in your case record supports such a finding.

G. How do we use the functional criteria in 7.18?

1. When we use the functional criteria in 7.18, we consider all relevant information in your case record to determine the impact of your hematological disorder on your ability to function independently, appropriately, effectively, and on a sustained basis in a work setting. Factors we will consider when we evaluate your functioning under 7.18 include, but are not limited to: your symptoms, the frequency and duration of complications of your hematological disorder, periods of exacerbation and remission, and the functional impact of your treatment, including the side effects of your medication.

2. Repeated complications means that the complications occur on an average of three times a year, or once every 4 months, each lasting 2 weeks or more; or the complications do not last for 2 weeks but occur substantially more frequently than three times in a year or once every 4 months; or they occur less frequently than an average of three times a year or once every 4 months but last substantially longer than 2 weeks. Your impairment will satisfy this criterion regardless of whether you have the same kind

of complication repeatedly, all different complications, or any other combination of complications; for example, two of the same kind of complication and a different one. You must have the required number of complications with the frequency and duration required in this section. Additionally, the complications must occur within the period we are considering in connection with your application or continuing disability review.

3. To satisfy the functional criteria in 7.18, your hematological disorder must result in a “marked” level of limitation in one of three general areas of functioning: activities of daily living, social functioning, or difficulties in completing tasks due to deficiencies in concentration, persistence, or pace. Functional limitations may result from the impact of the disease process itself on your mental functioning, physical functioning, or both your mental and physical functioning. This limitation could result from persistent or intermittent symptoms, such as pain, severe fatigue, or malaise, resulting in a limitation of your ability to do a task, to concentrate, to persevere at a task, or to perform the task at an acceptable rate of speed. (Severe fatigue means a frequent sense of exhaustion that results in significant reduced physical activity or mental function. Malaise means frequent feelings of illness, bodily discomfort, or lack of well-being that result in significantly reduced physical activity or mental function.) You may also have limitations because of your treatment and its side effects.

4. Marked limitation means that the symptoms and signs of your hematological disorder interfere seriously with your ability to function. Although we do not require the use of such a scale, “marked” would be the fourth point on a five-point scale consisting

of no limitation, mild limitation, moderate limitation, marked limitation, and extreme limitation. We do not define “marked” by a specific number of different activities of daily living or different behaviors in which your social functioning is impaired, or a specific number of tasks that you are able to complete, but by the nature and overall degree of interference with your functioning. You may have a marked limitation when several activities or functions are impaired, or even when only one is impaired. Additionally, you need not be totally precluded from performing an activity to have a marked limitation, as long as the degree of limitation interferes seriously with your ability to function independently, appropriately, and effectively. The term “marked” does not imply that you must be confined to bed, hospitalized, or in a nursing home.

5. Activities of daily living include, but are not limited to, such activities as doing household chores, grooming and hygiene, using a post office, taking public transportation, or paying bills. We will find that you have a “marked” limitation in activities of daily living if you have a serious limitation in your ability to maintain a household or take public transportation because of symptoms such as pain, severe fatigue, anxiety, or difficulty concentrating, caused by your hematological disorder (including complications of the disorder) or its treatment, even if you are able to perform some self-care activities.

6. Social functioning includes the capacity to interact with others independently, appropriately, effectively, and on a sustained basis. It includes the ability to communicate effectively with others. We will find that you have a “marked” limitation in maintaining

social functioning if you have a serious limitation in social interaction on a sustained basis because of symptoms such as pain, severe fatigue, anxiety, or difficulty concentrating, or a pattern of exacerbation and remission, caused by your hematological disorder (including complications of the disorder) or its treatment, even if you are able to communicate with close friends or relatives.

7. Completing tasks in a timely manner involves the ability to sustain concentration, persistence, or pace to permit timely completion of tasks commonly found in work settings. We will find that you have a “marked” limitation in completing tasks if you have a serious limitation in your ability to sustain concentration or pace adequate to complete work-related tasks because of symptoms, such as pain, severe fatigue, anxiety, or difficulty concentrating caused by your hematological disorder (including complications of the disorder) or its treatment, even if you are able to do some routine activities of daily living.

H. How do we consider your symptoms, including your pain, severe fatigue, and malaise? Your symptoms, including pain, severe fatigue, and malaise, may be important factors in our determination whether your hematological disorder(s) meets or medically equals a listing, or in our determination whether you are otherwise able to work. We cannot consider your symptoms unless you have medical signs or laboratory findings showing the existence of a medically determinable impairment(s) that could reasonably be expected to produce the symptoms. If you have such an impairment(s), we will evaluate the intensity, persistence, and functional effects of your symptoms using the

rules throughout 7.00 and in our other regulations. (See sections 404.1528, 404.1529, 416.928, and 416.929 of this chapter.) Additionally, when we assess the credibility of your complaints about your symptoms and their functional effects, we will not draw any inferences from the fact that you do not receive treatment or that you are not following treatment without considering all of the relevant evidence in your case record, including any explanations you provide that may explain why you are not receiving or following treatment.

I. How do we evaluate episodic events in hematological disorders? Some of the listings in this body system require a specific number of events within a consecutive 12-month period. (See 7.05, 7.08, and 7.10A.) When we use such criteria, a consecutive 12-month period means a period of 12 consecutive months, all or part of which must occur within the period we are considering in connection with your application or continuing disability review. These events must occur at least 30 days apart to ensure that we are evaluating separate events.

J. How do we evaluate hematological disorders that do not meet one of these listings?

1. These listings are only common examples of hematological disorders that we consider severe enough to prevent a person from doing any gainful activity. If your disorder does not meet the criteria of any of these listings, we must consider whether you have a disorder that satisfies the criteria of a listing in another body system. For example,

we will evaluate hemophilic joint deformity or bone or joint pain from myelofibrosis under 1.00; polycythemia vera under 3.00, 4.00, or 11.00; chronic iron overload resulting from repeated RBC transfusion (transfusion hemosiderosis) under 3.00, 4.00, or 5.00; and the effects of intracranial bleeding or stroke under 11.00 or 12.00.

2. If you have a severe medically determinable impairment(s) that does not meet a listing, we will determine whether your impairment(s) medically equals a listing. (See sections 404.1526 and 416.926 of this chapter.) Hematological disorders may be associated with disorders in other body systems, and we consider the combined effects of multiple impairments when we determine whether they medically equal a listing. If your impairment(s) does not medically equal a listing, you may or may not have the residual functional capacity to engage in substantial gainful activity. We proceed to the fourth, and, if necessary, the fifth steps of the sequential evaluation process in sections 404.1520 and 416.920. We use the rules in sections 404.1594, 416.994, and 416.994a of this chapter, as appropriate, when we decide whether you continue to be disabled.

7.01 Category of Impairments, Hematological Disorders

7.05 Hemolytic anemias, including sickle cell disease, thalassemia, and their variants (see 7.00C), with:

A. Documented painful (vaso-occlusive) crises requiring parenteral (intravenous or intramuscular) narcotic medication, occurring at least six times within a 12-month

period with at least 30 days between crises.

OR

B. Complications of hemolytic anemia requiring at least three hospitalizations within a 12-month period and occurring at least 30 days apart. Each hospitalization must last at least 48 hours, which can include hours in a hospital emergency department or comprehensive sickle cell disease center immediately before the hospitalization (see 7.00C2).

OR

C. Hemoglobin measurements of 7.0 grams per deciliter (g/dL) or less, occurring at least three times within a 12-month period with at least 30 days between measurements.

OR

D. Beta thalassemia major requiring life-long RBC transfusions at least once every 6 weeks to maintain life (see 7.00C4).

7.08 Disorders of thrombosis and hemostasis, including hemophilia and thrombocytopenia (see 7.00D), with complications requiring at least three hospitalizations within a 12-month period and occurring at least 30 days apart. Each

hospitalization must last at least 48 hours, which can include hours in a hospital emergency department or comprehensive hemophilia treatment center immediately before the hospitalization (see 7.00D2).

7.10 Disorders of bone marrow failure, including myelodysplastic syndromes, aplastic anemia, granulocytopenia, and myelofibrosis (see 7.00E), with:

A. Complications of bone marrow failure requiring at least three hospitalizations within a 12-month period and occurring at least 30 days apart. Each hospitalization must last at least 48 hours, which can include hours in a hospital emergency department immediately before the hospitalization (see 7.00E2).

OR

B. Myelodysplastic syndromes or aplastic anemias requiring life-long RBC transfusions at least once every 6 weeks to maintain life (see 7.00E3).

7.17 Hematological disorders treated by bone marrow or stem cell transplantation (see 7.00F). Consider under a disability for at least 12 consecutive months from the date of transplantation. After that, evaluate any residual impairment(s) under the criteria for the affected body system.

7.18 Repeated complications of hematological disorders (see 7.00G2), including

those complications listed in 7.05, 7.08, and 7.10 but without the requisite findings for those listings, or other complications (for example, anemia, osteonecrosis, retinopathy, skin ulcers, silent central nervous system infarction, cognitive or other mental limitation, or limitation of joint movement), resulting in significant, documented symptoms or signs (for example, pain, severe fatigue, malaise, fever, night sweats, headaches, joint or muscle swelling, or shortness of breath), and one of the following at the marked level (see 7.00G4):

A. Limitation of activities of daily living (see 7.00G5).

B. Limitation in maintaining social functioning (see 7.00G6).

C. Limitation in completing tasks in a timely manner due to deficiencies in concentration, persistence, or pace (see 7.00G7).

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13.00 MALIGNANT NEOPLASTIC DISEASES

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K. How do we evaluate specific malignant neoplastic diseases?

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2. Leukemia.

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c. Chronic lymphocytic leukemia.

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ii. We evaluate the complications and residual impairment(s) from chronic lymphocytic leukemia (CLL) under the appropriate listings, such as 13.05A2 or an appropriate listing in 7.00.

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3. Macroglobulinemia or heavy chain disease. * * * We evaluate the resulting impairment(s) under the criteria of 7.00 or any other affected body system.

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Part B

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107.00 HEMATOLOGICAL DISORDERS

A. What hematological disorders do we evaluate under these listings?

1. We evaluate non-malignant (non-cancerous) hematological disorders, such as hemolytic anemias (107.05), disorders of thrombosis and hemostasis (107.08), and disorders of bone marrow failure (107.10). These disorders disrupt the normal development and function of white blood cells, red blood cells, platelets, and clotting-factor proteins (factors).

2. We evaluate malignant (cancerous) hematological disorders, such as lymphoma, leukemia, and multiple myeloma under the appropriate listings in 113.00, except for lymphoma associated with human immunodeficiency virus (HIV) infection, which we evaluate under 114.08E.

B. What evidence do we need to document that you have a hematological disorder? We need the following evidence to document that you have a hematological disorder:

1. A laboratory report of a definitive test that establishes a hematological disorder, signed by a physician; or

2. A laboratory report of a definitive test that establishes a hematological disorder that is not signed by a physician and a report from a physician that states you have the disorder; or

3. When we do not have a laboratory report of a definitive test, a persuasive report from a physician that a diagnosis of your hematological disorder was confirmed by appropriate laboratory analysis or other diagnostic method(s). To be persuasive, this report must state that you had the appropriate definitive laboratory test or tests for diagnosing your disorder and provide the results, or explain how your diagnosis was established by other diagnostic method(s) consistent with the prevailing state of medical knowledge and clinical practice.

4. We will make every reasonable effort to obtain the results of appropriate laboratory testing you have had. We will not purchase complex, costly, or invasive tests, such as tests of clotting-factor proteins, and bone marrow aspirations.

C. What are hemolytic anemias, and how do we evaluate them under 107.05?

1. Hemolytic anemias, both congenital and acquired, are disorders that result in premature destruction of red blood cells (RBCs). Hemolytic anemias include abnormalities of hemoglobin structure (hemoglobinopathies), abnormal RBC enzyme content and function, and RBC membrane (envelope) defects that are congenital or

acquired. The diagnosis of hemolytic anemia is based on hemoglobin electrophoresis or analysis of the contents of the RBC (enzymes) and membrane. Examples of congenital hemolytic anemias include sickle cell disease, thalassemia, and their variants, and hereditary spherocytosis. Acquired hemolytic anemias may result from autoimmune disease (for example, systemic lupus erythematosus) or mechanical devices (for example, heart valves, intravascular patches).

2. The hospitalizations in 107.05B do not all have to be for the same complication of the hemolytic anemia. They may be for three different complications of the disorder. Examples of complications of hemolytic anemia that may result in hospitalization include dactylitis, osteomyelitis, painful (vaso-occlusive) crisis, pulmonary infections or infarctions, acute chest syndrome, pulmonary hypertension, chronic heart failure, gallbladder disease, hepatic (liver) failure, renal (kidney) failure, nephrotic syndrome, aplastic crisis, and strokes. We will count the hours you receive emergency treatment in a comprehensive sickle cell disease center immediately before the hospitalization if this treatment is comparable to the treatment provided in a hospital emergency department.

3. For 107.05C, we do not require hemoglobin to be measured during a period in which you are free of pain or other symptoms of your disorder. We will accept hemoglobin measurements made while you are experiencing complications of your hemolytic anemia.

4. 107.05D refers to the most serious type of beta thalassemia major in which the

bone marrow cannot produce sufficient numbers of normal RBCs to maintain life. The only available treatments for beta thalassemia major are life-long RBC transfusions (sometimes called hypertransfusion) or bone marrow transplantation. For purposes of 107.05D, we do not consider prophylactic RBC transfusions to prevent strokes or other complications in sickle cell disease and its variants to be of equal significance to life-saving RBC transfusions for beta thalassemia major. However, we will consider the functional limitations associated with prophylactic RBC transfusions and any associated side effects (for example, iron overload) under functional equivalence and any affected body system(s). We will also evaluate strokes and resulting complications under 111.00 and 112.00.

D. What are disorders of thrombosis and hemostasis, and how do we evaluate them under 107.08?

1. Disorders of thrombosis and hemostasis include both clotting and bleeding disorders, and may be congenital or acquired. These disorders are characterized by abnormalities in blood clotting that result in hypercoagulation (excessive blood clotting) or hypocoagulation (inadequate blood clotting). The diagnosis of a thrombosis or hemostasis disorder is based on evaluation of plasma clotting-factor proteins (factors) and platelets. Protein C or protein S deficiency and Factor V Leiden are examples of hypercoagulation disorders. Hemophilia, von Willebrand disease, and thrombocytopenia are examples of hypocoagulation disorders. Acquired excessive blood clotting may result from blood protein defects and acquired inadequate blood clotting (for example, acquired

hemophilia A) may be associated with inhibitor autoantibodies.

2. The hospitalizations in 107.08 do not all have to be for the same complication of a disorder of thrombosis and hemostasis. They may be for three different complications of the disorder. Examples of complications that may result in hospitalization include anemias, thromboses, embolisms, and uncontrolled bleeding requiring multiple factor concentrate infusions or platelet transfusions. We will also consider any surgery that you have, even if it is not related to your hematological disorder, to be a complication of your disorder of thrombosis and hemostasis if you require treatment with clotting-factor proteins (for example, factor VIII or IX) or anticoagulant medication to control bleeding or coagulation in connection with your surgery. We will count the hours you receive emergency treatment in a comprehensive hemophilia treatment center immediately before the hospitalization if this treatment is comparable to the treatment provided in a hospital emergency department.

E. What are disorders of bone marrow failure, and how do we evaluate them under 107.10?

1. Disorders of bone marrow failure may be congenital or acquired, characterized by bone marrow that does not make enough healthy RBCs, platelets, or granulocytes (specialized types of white blood cells); there may also be a combined failure of these bone marrow-producing cells. The diagnosis is based on peripheral blood smears and bone marrow aspiration or bone marrow biopsy, but not peripheral blood smears alone.

Examples of these disorders are myelodysplastic syndromes, aplastic anemia, granulocytopenia, and myelofibrosis. Acquired disorders of bone marrow failure may result from viral infections, chemical exposure, or immunologic disorders.

2. The hospitalizations in 107.10A do not all have to be for the same complication of bone marrow failure. They may be for three different complications of the disorder. Examples of complications that may result in hospitalization include uncontrolled bleeding, anemia, and systemic bacterial, viral, or fungal infections.

3. For 107.10B, the requirement of life-long RBC transfusions to maintain life in myelodysplastic syndromes or aplastic anemias has the same meaning as it does for beta thalassemia major. (See 107.00C4.)

F. How do we evaluate bone marrow or stem cell transplantation under 107.17?

We will consider you to be disabled for 12 months from the date of bone marrow or stem cell transplantation, or we may consider you to be disabled for a longer period if you are experiencing any serious post-transplantation complications, such as graft-versus-host (GVH) disease, frequent infections after immunosuppressive therapy, or significant deterioration of organ systems. We do not restrict our determination of the onset of disability to the date of the transplantation in 107.17. We may establish an earlier onset of disability due to your transplantation if evidence in your case record supports such a finding.

G. How do we consider your symptoms, including your pain, severe fatigue, and malaise? Your symptoms, including pain, severe fatigue, and malaise, may be important factors in our determination whether your hematological disorder meets or medically equals a listing, or in our determination whether you otherwise have marked and severe functional limitations. We cannot consider your symptoms unless you have medical signs or laboratory findings showing the existence of a medically determinable impairment(s) that could reasonably be expected to produce the symptoms. If you have such an impairment(s), we will evaluate the intensity, persistence, and functional effects of your symptoms using the rules throughout 107.00 and in our other regulations. (See sections 416.928 and 416.929 of this chapter.) Additionally, when we assess the credibility of your complaints about your symptoms and their functional effects, we will not draw any inferences from the fact that you do not receive treatment or that you are not following treatment without considering all of the relevant evidence in your case record, including any explanations you provide on why you are not receiving or following treatment.

H. How do we evaluate episodic events in hematological disorders? Some of the listings in this body system require a specific number of events within a consecutive 12-month period. (See 107.05, 107.08, and 107.10A.) When we use such criteria, a consecutive 12-month period means a period of 12 consecutive months, all or part of which must occur within the period we are considering in connection with your application or continuing disability review. These events must occur at least 30 days apart to ensure that we are evaluating separate events.

I. How do we evaluate hematological disorders that do not meet one of these listings?

1. These listings are only common examples of hematological disorders that we consider severe enough to result in marked and severe functional limitations. If your disorder does not meet the criteria of any of these listings, we must consider whether you have a disorder that satisfies the criteria of a listing in another body system. For example, we will evaluate hemophilic joint deformity under 101.00; polycythemia vera under 103.00, 104.00, or 111.00; chronic iron overload resulting from repeated RBC transfusion (transfusion hemosiderosis) under 103.00, 104.00, or 105.00; and the effects of intracranial bleeding or stroke under 111.00 or 112.00.

2. If you have a severe medically determinable impairment(s) that does not meet a listing, we will determine whether your impairment(s) medically equals a listing. (See section 416.926 of this chapter.) Hematological disorders may be associated with disorders in other body systems, and we consider the combined effects of multiple impairments when we determine whether they medically equal a listing. If your impairment(s) does not medically equal a listing, we will also consider whether it functionally equals the listings. (See section 416.926a of this chapter.) We use the rules in §416.994a of this chapter when we decide whether you continue to be disabled.

107.01 Category of Impairments, Hematological Disorders

107.05 Hemolytic anemias, including sickle cell disease, thalassemia, and their variants (see 107.00C), with:

A. Documented painful (vaso-occlusive) crises requiring parenteral (intravenous or intramuscular) narcotic medication, occurring at least six times within a 12-month period with at least 30 days between crises.

OR

B. Complications of hemolytic anemia requiring at least three hospitalizations within a 12-month period and occurring at least 30 days apart. Each hospitalization must last at least 48 hours, which can include hours in a hospital emergency department or comprehensive sickle cell disease center immediately before the hospitalization (see 107.00C2).

OR

C. Hemoglobin measurements of 7.0 grams per deciliter (g/dL) or less, occurring at least three times within a 12-month period with at least 30 days between measurements.

OR

D. Beta thalassemia major requiring life-long RBC transfusions at least once

every 6 weeks to maintain life (see 107.00C4).

107.08 Disorders of thrombosis and hemostasis, including hemophilia and thrombocytopenia (see 107.00D), with complications requiring at least three hospitalizations within a 12-month period and occurring at least 30 days apart. Each hospitalization must last at least 48 hours, which can include hours in a hospital emergency department or comprehensive hemophilia treatment center immediately before the hospitalization (see 107.00D2).

107.10 Disorders of bone marrow failure, including myelodysplastic syndromes, aplastic anemia, granulocytopenia, and myelofibrosis (see 107.00E), with:

A. Complications of bone marrow failure requiring at least three hospitalizations within a 12-month period and occurring at least 30 days apart. Each hospitalization must last at least 48 hours, which can include hours in a hospital emergency department immediately before the hospitalization (see 107.00E2).

OR

B. Myelodysplastic syndromes or aplastic anemias requiring life-long RBC transfusions at least once every 6 weeks to maintain life (see 107.00E3).

107.17 Hematological disorders treated by bone marrow or stem cell

transplantation (see 107.00F). Consider under a disability for at least 12 consecutive months from the date of transplantation. After that, evaluate any residual impairment(s) under the criteria for the affected body system.

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